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A case of cardiac sarcoidosis with successful heart transplantation after COVID-19 infection

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Abstract

Arrhythmogenic right ventricular cardiomyopathy and cardiac sarcoidosis can both present with ventricular tachycardia. We report a case of a patient whose histological diagnosis was not only confirmed by the transplanted heart but who also underwent successful transplantation after overcoming COVID-19.

Learning objectives

Similarities in the clinical presentation of cardiac sarcoidosis (CS) and arrhythmogenic right ventricular cardiomyopathy (ARVC) Management differences between CS and ARVC Successful heart transplantation after COVID-19

Introduction

The COVID-19 pandemic has led to a wide range of challenges in the health care system. As of February 2021, a total of 113 144 824 people had been affected, leading to 2 510 343 fatalities worldwide [1]. Most of the scientific focus has been directed toward describing the virus, its clinical course, and treatment options. However, there are limited data on how COVID-19 affects solid organ transplant patients. There are case reports describing heart transplant recipients who acquired COVID-19, but there are not many anecdotal cases where a successful heart transplant was performed in a patient who recovered from COVID-19. A wide range of differential exits for ventricular tachycardia. Two rare etiologies of ventricular arrhythmias are cardiac sarcoidosis (CS) and arrhythmogenic right ventricular cardiomyopathy (ARVC).

Sarcoidosis is characterized by the presence of non-necrotizing granulomas that involve multiple organs, most commonly the lungs and lymph nodes. CS is clinically manifested in about 5% of patients with

systemic sarcoidosis, but more than 25% may have evidence of cardiac involvement on autopsy or imaging studies [2]. There are no specific features of myocardial involvement pathognomonic for CS, which makes the diagnosis a challenging one. We present the case of a patient with post-cardiac arrest and concomitant COVID-19 infection who was initially diagnosed with ARVC, but a biopsy of the explanted heart revealed CS.

Case Report

A healthy 37-year-old female with no past medical history suffered sudden cardiac arrest at home shortly after testing positive for COVID-19. She was found to be in ventricular fibrillation (VF), and return to spontaneous circulation was achieved approximately 1 hour after the arrest. Imaging studies demonstrated extensive consolidation bilaterally suggestive of severe COVID-19 pneumonia/acute respiratory distress syndrome, therefore her cardiac arrest was thought to be secondary to hypoxia. Subsequently, the patient recovered, and a secondary prevention implantable cardioverter defibrillator (ICD) was implanted before discharge.

A few days later, she presented to another hospital with syncope and recurrent ventricular arrhythmias treated by multiple appropriate ICD shocks. ICD interrogation showed 2 ventricular tachycardia (VT) episodes, 20 VF episodes, and a total of 19 40-J shocks delivered, all successful at terminating the arrhythmias. Left heart catheterization showed normal coronaries and left ventricular ejection fraction (LVEF) of 40%. She was taken to the electrophysiology laboratory for an attempt at mapping and ablation of VT which could not be performed successfully due to recurrent episodes of intra-operative VF requiring multiple shocks. The right ventricular endocardial bipolar voltage maps obtained were suggestive of ARVC (Fig. 1), vs. infiltrative myocarditis. After a failed attempt at VT ablation, the patient was transferred to our institution intubated, sedated, on vasopressor support and IV antiarrhythmic, for possible heart transplant evaluation in the context of ventricular electrical storm.

While admitted to our hospital, repeat imaging studies demonstrated multiple nodules throughout the right lower lobe (Fig. 2A and B). An echocardiogram showed LVEF of 35% and moderately decreased function of the RV (Fig. 2C). The patient was managed with amiodarone and lidocaine infusions. Her course was further complicated by cardiac tamponade requiring a pericardial window. With a tentative diagnosis of the ARVC complicated by refractory life-threatening arrhythmias not amenable to mapping/ablation, a heart transplant evaluation was started and the patient was successfully listed as status 3 by exception. Her episodes of arrhythmia improved, and she was extubated and weaned off lidocaine infusion. The patient's scattered airspace consolidations on repeat images were attributed to resolving COVID-19 pneumonia. She underwent a successful heart transplant 2 months after the initial presentation. The explanted heart showed extensive non-necrotizing granulomatous inflammation and fibrosis diagnostic of CS (Fig. 3). The patient had an uneventful post-transplant course and was discharged on immunosuppressive therapy. She was referred for a positron emission tomography (PET) scan to look for extra-cardiac disease.

Discussion

ARVC and CS can both present with RV dilation and VT. CS presenting as ARVC has been reported previously in the literature [3, 4]. Some patients with CS can meet the criteria for ARVC, which can be explained by the involvement of the right ventricle in both entities. ARVC is characterized by fibrous/fatty infiltration of the right ventricle. The patient typically presents with syncope, palpitations, or cardiac arrest. The diagnosis of ARVC is established by meeting the 2010 International Task Force Criteria [5]. In our case, the patient had met possible 2010 International Task Force Criteria with two minor criteria including inverted T waves in leads V1, V2, V3, and V4 in the presence of complete right bundle branch block (Fig. 2D) and >500 ventricular extrasystoles per 24 hours. The clinical presentation

of cardiac arrest, recurrent arrhythmia, and findings of RV scarring in voltage mapping led us to believe the working diagnosis for our patient was ARVC. There are no specific treatment options for ARVC besides antiarrhythmic medications and ICD for secondary prevention [6].

CS also has patchy involvement of the heart muscle. It classically presents with atrioventricular conduction disease, arrhythmias, and heart failure [2]. At times, isolated RV sarcoidosis can present with life-threatening arrhythmias that mimic ARVC. CS is usually a pathologic diagnosis, but endomyocardial biopsy is rarely done to make the diagnosis. Cardiac PET is the preferred image for diagnosis and monitoring treatment response since it is both sensitive and specific [6]. In our patient, the presence of an ICD, as well as frequent hemodynamic instability, precluded us from obtaining a cardiac magnetic resonance image or cardiac PET computed tomography. The diagnostic distinction between ARVC and CS is important since both are treated differently. CS is treated with immunosuppression consisting of high-dose steroids initially and then switching or adding a steroid-sparing agent [6].

Furthermore, the COVID-19 pandemic has added an extra layer of complexity when differentiating between diseases that may have similar presentations such as CS and ARVC. As previously mentioned, COVID-19 can have a long-standing effect on the lung and cardiovascular systems. The pulmonary infiltrates present in our patient were attributed to the underlying viral infection, which further obscured the diagnosis of CS. Also, there have been increasing reports of post-COVID patients developing postural orthostatic tachycardia syndrome. The increase in the number of patients affected by COVID-19 is inevitable; the implication of COVID-19 positivity in the pre-transplant patient will need to be considered in transplant centers. This case highlights the importance of tissue to unmask CS since the examination of the explanted heart was critical to making the final diagnosis. It also depicts a case of a patient who underwent successful heart transplantation after overcoming COVID-19 pneumonia.

Acknowledgments

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Conflict of Interest

There is no conflict of interest

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Legends

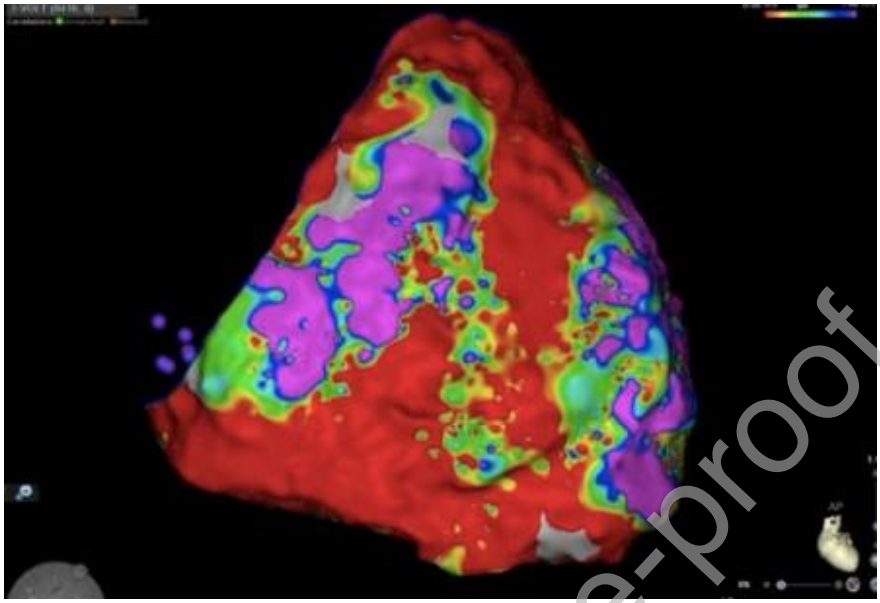

Figure 1 Abnormal bipolar endocardial voltage map of the right ventricle showing areas of scarring, a substrate pattern commonly seen in arrhythmogenic right ventricular cardiomyopathy. Right anterior oblique view.

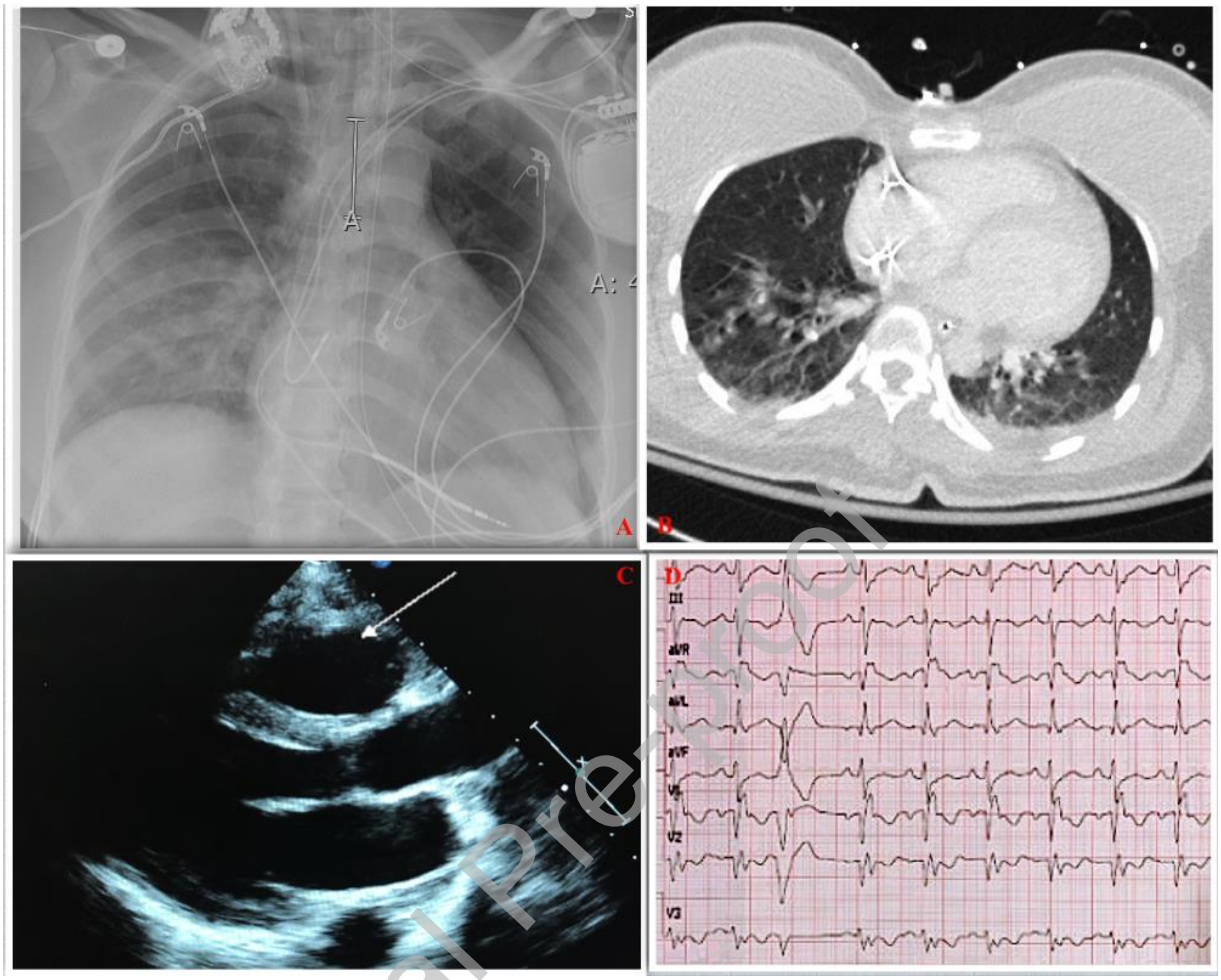

Figure 2 (A) Chest X-ray image showing mild pulmonary venous congestion with interstitial opacities in the right perihilar and right lower lobe. (B) Computed tomography of the chest notable for multiple nodules throughout the right lower lobe. (C) Parasternal long-axis view. Arrow points to dilated right ventricle. (D) Inverted T waves in leads V1, V2, V3, and V4 in the presence of complete right bundle branch block.

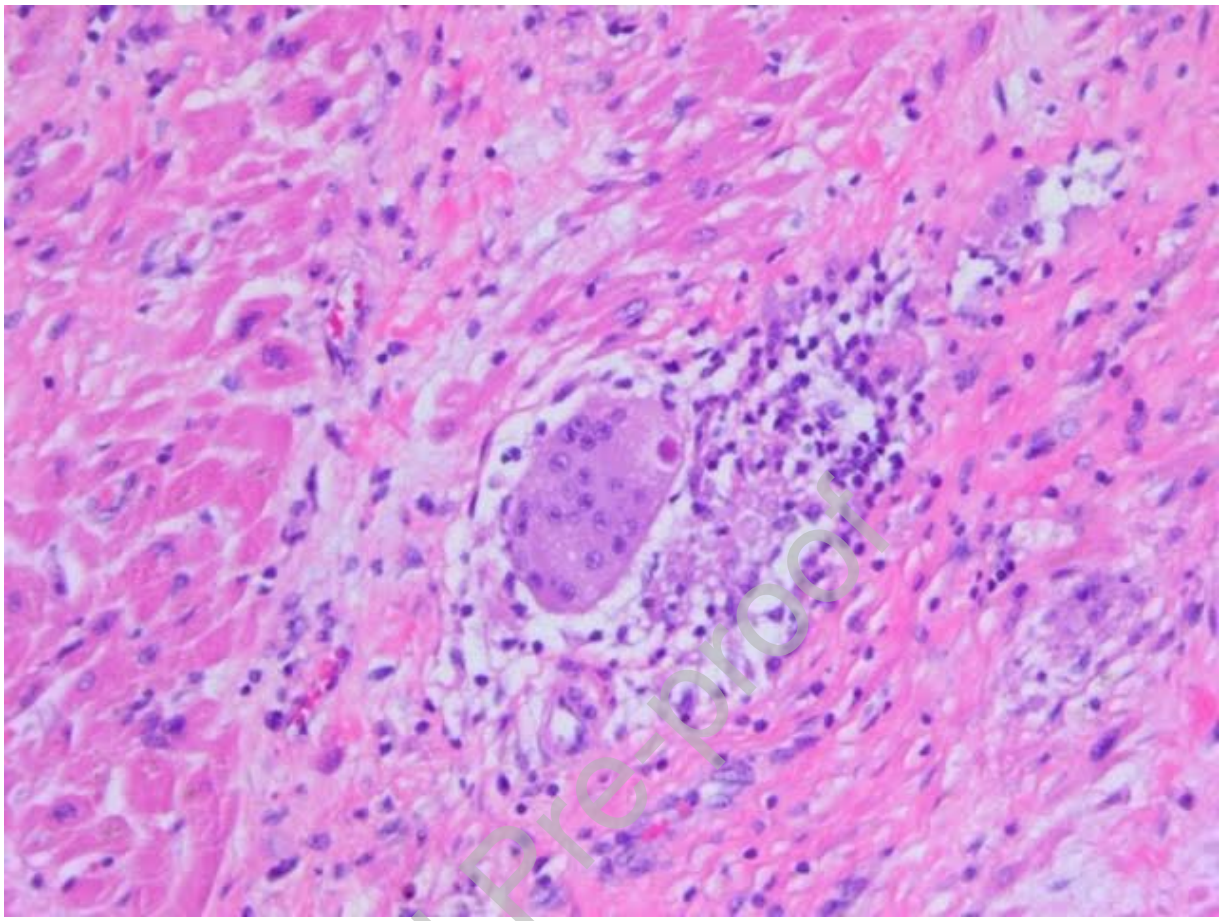

Figure 3 Myocardium of the explanted heart showing compact non-necrotizing granuloma surrounded by a cuff of lymphocytes and associated with extensive fibrosis characteristic of sarcoidosis. Hematoxylin and eosin; 200X.